<Clinical Studies>A Case of Liposarcoma Originating in the Chest Wall

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A Case of Liposarcoma Originating in the Chest Wall

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Abstract

We encountered and reported one such rare case of liposarcoma which originated in the chest wall. A 60-year-old man came to our hospital with the chief complaint of a phyma in the right anterior chest wall. On palpation, a hard and non-mobile phyma measuring 3 × 3 cm was felt in the chest wall. Chest CT showed a phyma measuring 2.2 × 1.5 cm in the right anterior chest. The periphery of the phyma was smooth, and had a well-defined boundary with the surrounding tissues. Ultrasonic examination revealed that the tumor existed between the major and minor pectoral muscles. The inside of the tumor was nearly uniform, and showed low echo. Punctured cytological examination revealed scattered atypical cells with spindle, foamy or vacuolar sporophores on the mucoid matrix. A fat staining examination revealed lipoblasts with oil red-positive granules. Based on these findings, the patient was diagnosed as having myxoid type liposarcoma. Operation consisted of resection of the skin, subcutaneous tissues, mammary gland, part of major and minor pectoral muscles, the fourth and fifth ribs and pleura. The Reconstruction of the chest wall was performed for defects in the ribs and pleura using Marlex Mesh. Histopathological findings revealed that the tumor was myxoid type liposarcoma.

Introduction

Liposarcoma occurs relatively frequently among various types of maligmant tissue tumors; its incidence is the third highest after malignant fibrous histiocytoma and rhabdomyosarcoma[1,2]. This disease mostly occurs in the lower extremity and the retroperitoneum, and it rarely develops in the chest wall[3,4]. We encountered one such rare case of liposarcoma which originated in the chest wall. A report on this is presented here with literature review.

Key words: Liposarcoma, Chest wall
索引用語: 脂肪肉腫，胸壁
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Case presentation

A 60-year-old man came to the outpatient clinic of the First Department of Surgery, Juntendo University, School of Medicine, on March 1, 1995, with the chief complaint of a phyma in the right anterior chest. On palpation, a hard and non-mobile phyma measuring 3×3 cm was felt in the right anterior chest. No superficial lymph nodes, including axially lymph nodes, were felt.

Fig. 1 Chest CT shows a phyma measuring 2.2×1.5 cm in the right anterior chest wall.

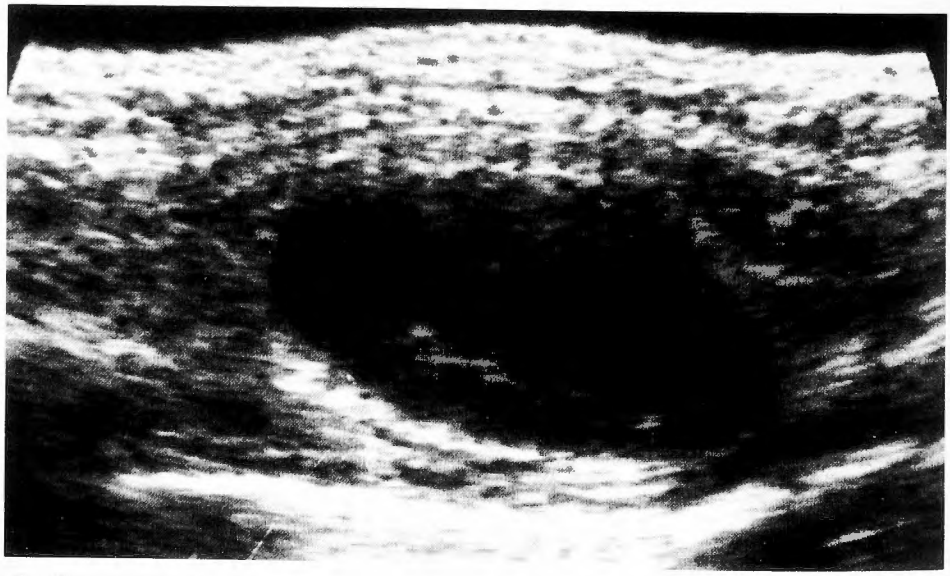

Fig. 2 Ultrasonic examination reveals that the tumor existed between the major and minor pectoral muscles.

The patient had subtotal gastrectomy and cholecystectomy because of early gastric cancer [m, IIc+III. tubl, ly_0, v_0, n_0] and cholecystolithiasis on February 2, 1994.

On March 8, 1995, the patient was admitted to the hospital. Chest CT showed a phyma meas-

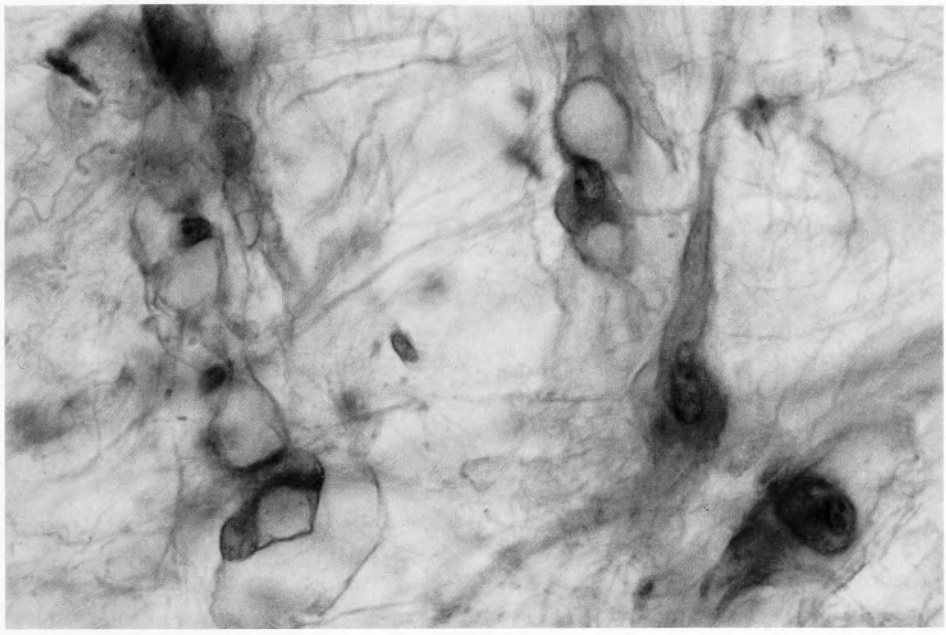

Fig. 3 Punctured cytological examination reveals scattered atypical cells with spindle, foamy or vacuolar sporophores with no connectivity, on the mucoid matrix.

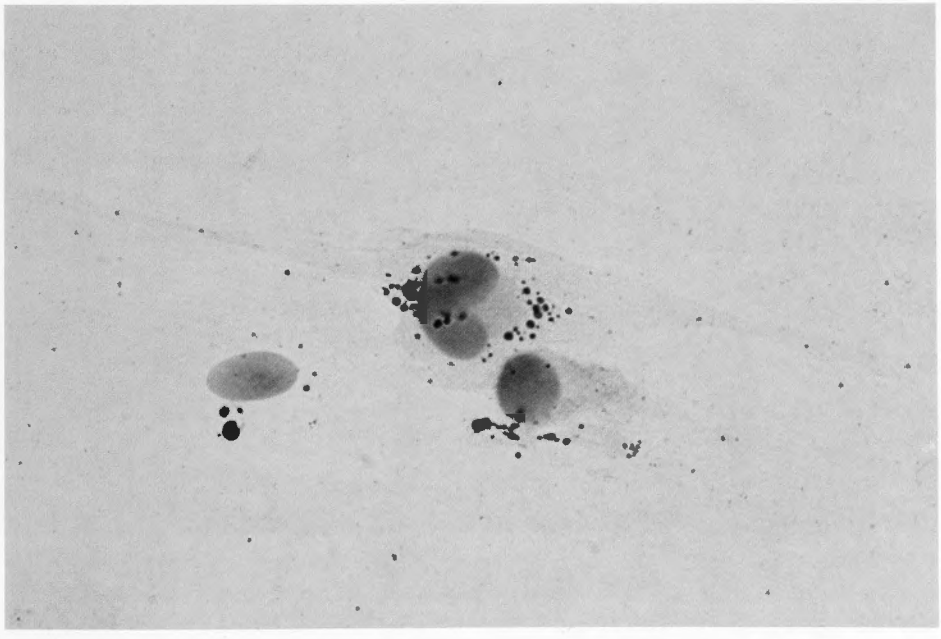

Fig. 4 A fat staining examination reveals lipoblasts with oil red-positive granules.

uring 2.2 × 1.5 cm in the right anterior chest (Fig. 1). The periphery of the phyma was smooth, and had a wall-defined boundary with the surrounding tissues. The phyma existed under the pectoral muscle and compressed the muscular tissues laterally. No findings suggesting the infiltration of the phyma into the muscular tissues or ribs were observed. The inside of the tumor was almost uniform with a low-absorption area where a Hounsfild unit was similar to that obtained in fatty tissues. This area was visualized faintly by contrasted CT.

Ultrasonic examination revealed that the tumor existed between the major and minor pectoral muscles (Fig. 2). The inside of the tumor was nearly uniform, and showed low echo.

Punctured cytological examination revealed scattered atypical cells with spindle, foamy or vacuolar sporophores, which were probably derived from non-epithelial tissues with no connectivity, on the mucoid matrix (Fig. 3). Matured lipocytes of various sizes with the nuclei compressed by the periphery and large vacuolar sporophores were occasionally found. The sporophores of tumor cells were vacuolated, and fat staining revealed lipoblasts with oil red-positive granules (Fig. 4). Based on these findings, the patient was diagnosed as having myxoid type liposarcoma.

A systemic gallium scintigraphy showed no abnormal accumulation of radioactive substances in the anterior chest. A systemic bone scintigraphy revealed no abnormal accumulation of radioactive substances in the ribs.

A biochemical test performed at hospitalization revealed no abnormal findings, except for a tumor marker CEA that slightly increased to 7.4 ng/ml.

On July 3, 1995, operation was performed. Operation consisted of resection of the skin, subcutaneous tissues, mammary gland, part of major and minor pectoral muscles, the fourth and fifth ribs and pleura. The Reconstruction of the chest wall was performed for defects in the ribs and pleura using Marlex Mesh.

The resected specimen was a yellowish-white and elastic tumor covered with a tunica, measur-

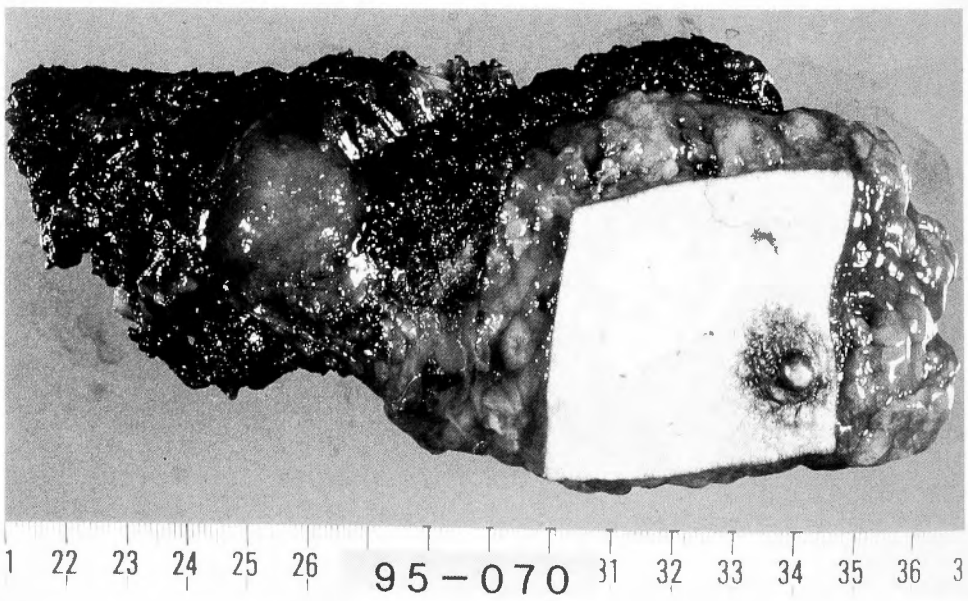

Fig. 5 The resected specimen shows a yellowish white and elastic tumor covered with a tunica, measuring 2.5 × 2.5 × 1.2 cm

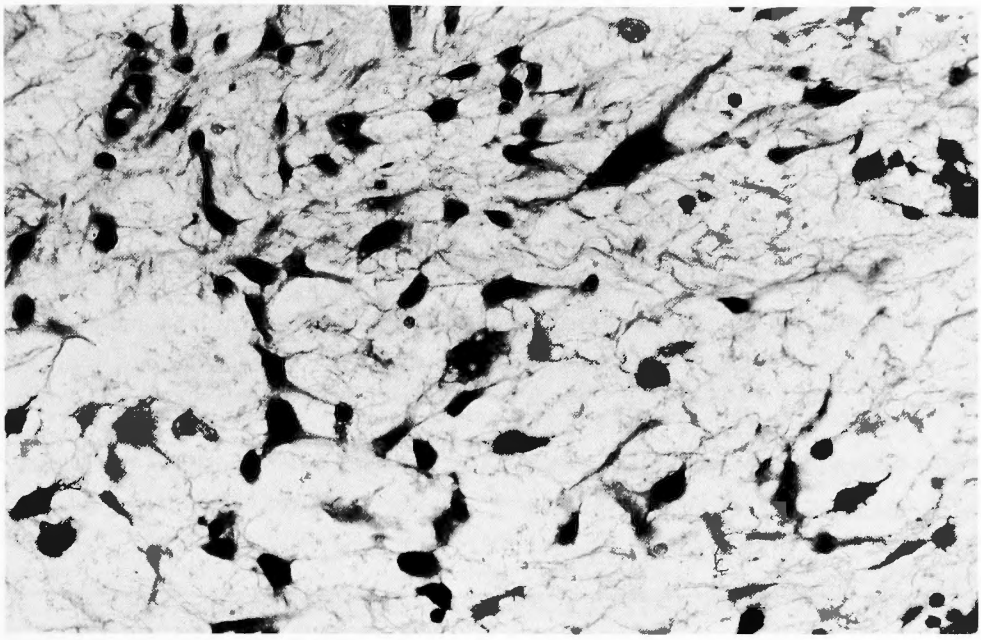

Fig. 6 Histopathological findings reveal a tumor was occupied by spindle or semicircular atypical cells and diagnosed mxoid type liposarcoma.

ing 2.5 × 2.5 × 1.2 cm (Fig. 5). It existed between major and minor pectaral muscles. Slight adhesion of the tumor to the surrounding tissues and pectoralis major muscle was observed.

Histopathological findings revealed the tumor occupied by spindle or semicircular atypical cells (Fig. 6). There were various sizes and variations of nuclei. The sporophores of tumor cells were vacuolated, and fat staining revealed lipoblasts with oil red-positive granules. Based on these findings, the tumor was considered as myxoid type liposarcoma. Tumor cells slightly infiltrated into the surrounding tissues and major pectoral muscle, but not into the vein or resected edge.

Discussion

Liposarcoma accounts for 15–20% of the cases of malignant soft tissue tumor, and its incidence is the third highest after malignant fibrous histiocytoma and rhabdomyosarcoma[1,2]. This disease occurs most frequently in the lower extremity, followed by the retroperitoneum and the trunk, and rarely develops in the chest wall[3,4]. The incidence of anatomic distribution of 136 liposarcomas was 54% for the lower limb and buttock, 21% for the retroperitoneum and mesentery, 10% for upper limb and shoulder and 7% for the trunk[5]. The averaged age of liposarcoma was 49.6 years and the raito of men to women was 1:0.86[5]. A search of the medical literature up to the present revealed that only 10 cases of liposarcoma originating in the chest wall, including our patient, have been reported in Japan[3,6,7] (Table 1). In the series of 10 cases of liposarcoma originating in the chest wall, the ratio of men to women was 5:4 except 1 case. The mean age at the time of diagnosis was 62.2 years. The mean age of liposarcoma originating in the chest wall was older than that of liposarcoma in other parts of the body. The size of tumors in the greatest diameter were from 2.0 cm to 21.5 cm and were more than 5 cm in most cases. The tumor experienced by the author was the smallest one

Table 1 Patients with liposarcoma originating in the chest wall reported in Japan (1985–1995)

No.	Author	Age	Sex	Size (cm)	Microscopic type	Operation	Prognosis
1	Takeuchi	—	—	—	—	WR	4 mo. alive
2	Hironaka	76	M	10×10	pleomorphic	none	2 mo. dead
3	Sato	39	F	5×3	myxoid	SR	dead (recurrence)
4	Hayashi	75	M	14×11	—	WR	24 mo. alive
5	Hatakeyama	62	M	10×8	pleomorphic	WR	16 mo. alive
6	Hayashi	41	F	5.5×3.0×1.5	—	WR	11 mo. alive
7	Kamata	70	M	18×14.2×8	mixed	SR	24 mo. alive
8	Oinuma	61	F	21.5×15×10.5	well diffe.	WR	10 mo. alive
9	Takahama	76	F	10×5×5	myxoid	WR	7 mo. alive
10	Our case	60	M	2.0×1.2	myxoid	WR	5 mo. alive

WR : wide resection SR : simple resection

in the reported cases, because the patient had been visiting the hospital periodically after operation for gastric cancer.

WHO's classification or regulations which have been set out to deal with malignant soft tissue tumors in Japan are used for the histopathological classification of liposarcoma. Based on these classifications, liposarcoma is classified into five types: 1) well differentiated, 2) myxoid, 3) round cell, 4) pleomorphic and 5) mixed[8,9]. In 10 cases of liposarcoma originating in the chest wall, the microscopic type was 3 myxoid type, 2 pleomorphic type, 1 mixed and well differentiated type and 3 miscellanous type. The myxoid type that our patient had occurs more frequently than other types of liposarcoma.

MRI and ultrasonic examinations are useful diagnostic tools. With CT examination, liposarcoma is characterized by a slightly higher Hounsfild unit than that of the normal fatty tissue and slightly un-uniform inside. However, it is often difficult to differentiate between liposarcoma and lipoma [10] Recent reports claim the usefulness of MRI examination for this purpose, and state that MRI examination could differentiate between liposarcoma and lipoma[11,12].

A histopathology, including a punctured cytological examination, is important for confirmed diagnosis. In our patient, confirmed diagnosis was obtained by preoperative punctured cytological examination.

The first option of treatment is the removal of lioposarcoma. Because liposarcoma is usually covered with a false tunica, it appears to be removed easily at a glance. Such a simple removal has a high rate of postoperative local recurrence[5,13]. Therefore, radical wide resection based on the concept of compartment was emphasized to prevent the recurrence of liposarcoma[14]. Moreover, it has been reported that curative wide resection that involves an en bloc resection with a tissue acting as a barrier placed around the tumor, provided favorable results[15]. Another suggested method is to remove the tumor as a mass by making an incision so that horizontally, the tumor is covered with a certain amount of normal tissue containing a living barrier, and longitudinally, by extending the

area removed to more than 5 cm from the boundary of the tumor[16]. A sufficiently extensive resection is also necessary to prevent local recurrence of a tumor involving the chest wall. However, definitive criteria for the range of removal have not been established. In 10 cases of liposarcoma originating in the chest wall, 7 cases underwent wide resection, 2 simple resection and 1 without operation. In 7 cases which underwent wide resection, recurrence was not recognized but one patient who underwent a simple resection, died of recurrence.

When more than three ribs are resected particularly in the anterior chest wall after the chest wall is excised together with other involved tissues, the mobility of the chest wall is inevitable and requires reconstruction. In our patient, the area removed was extended to 5 cm from the periphery of the tumor, and the skin, subcutaneous tissues, mammary gland, part of the major and minor pectoral muscles, the fourth and fifth ribs and pleura were resected together. In addition, the reconstruction of the chest wall was conducted for the chest wall defect using a Marlex Mesh. Favorable results were obtained.

Surgery was supplemented with chemotherapy and radiotherapy. Chemotherapy is systemic therapy which is intended to control invisible microfoci of metastases. In this sense, it is thought to be an effective supplementary therapy in preventing remote metastasis after radical operation. Adriamycin and Ifosfamide are used for single drug treatment, and CYVADIC therapy (cyclophosphamide, vincrisitin, adriamycin, and dacarbazine) is used for multidrug treatment. Successful cases have been reported[16,17]. Liposarcoma is thought to be relatively highly sensitive to radiation among various types of malignant soft tissue tumors. Radiotherapy has been used with operations intended to reduce the size of a tumor, and successful cases have been reported[17]. In the long run, however, it has various risk complications. Radiotherapy is commonly indicated in cases where remote metastasis is present before operation, for elderly patients, and patients who refuse resection [18,19]. Our patient received neither supplementary chemotherapy nor radiotherapy, because we considered that a successful radical operation was performed. Prognosis largely depends on the histologic type of liposarcoma. The five-year survival rate ranges from 20–30% for round cell and pleomorphic types to 80–90% for differentiated and myxoid types[13,20]. Generally, prognosis is the poorest in the pleomorphic type, followed by round cell, myxoid and differentiated types in this order. Therefore, the determination of a histologic type is clinically important. Based on the size of the tumor, the three-year survival rate is 88% for a size of 6 cm as compared to 50% for a size of 15 cm or larger according to literature[5]. Regardless of the histologic type, it has been reported that there were one of two recurrences in many cases at the time when 5 years have passed after the first operation[5]. On the other hand, it has been reported that radical operations had fewer recurrences[16].

The primary site of metastasis is the lung, followed by the intraperitoneal area and liver[5]. It appears necessary to follow up on our patient paying particular attention to remote metastasis.

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和文抄録

前胸壁原発脂肪肉腫の1例

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非常に稀な前胸壁原発脂肪肉腫の1例を経験したので報告する．症例は60歳，男性．右前胸壁腫瘤を主訴に来院した．右前胸部に弾性硬で大きさ 3×3 cm，可動性に乏しい腫瘤を触知した．胸部CT検査で右前胸壁に大きさ2.2×1.5 cm，表面平滑で辺縁明瞭な腫瘤を認めた．超音波検査で右胸壁大小胸筋間に内部ほぼ均一な低エコーを示す腫瘤を認めた．穿刺吸引細胞診で粘液様基質のなかに紡錘型または星状を呈する異型の強い細胞を認めた．脂肪染色のオイルレッド染色が陽性の脂肪芽細胞を認め，粘液型脂肪肉腫と診断した．手術は皮膚，皮下組織，乳腺，大小胸筋の一部および右第4・5肋骨を含め腫瘤を切除した．胸壁欠損部はmarlex mesh を用いて再建術を行った．病理組織学的所見は粘液型脂肪肉腫であった．